



Pulmonary Hypertension

Quick Reference Guide

2026

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1 CLINICAL SNAPSHOT

Definition: Pulmonary hypertension (PH) is a hemodynamic condition defined by a **mean pulmonary artery pressure (mPAP) >20 mm Hg** measured by right heart catheterization (RHC) at rest, reflecting elevated pressures in the pulmonary vasculature that progressively overload the right ventricle. PH encompasses five distinct WHO clinical groups — pulmonary arterial hypertension (PAH, Group 1), PH due to left heart disease (Group 2), PH due to lung diseases and hypoxia (Group 3), chronic thromboembolic PH (CTEPH, Group 4), and PH with unclear or multifactorial mechanisms (Group 5).

Accurate WHO group classification is a clinical and documentation requirement, as distinguishing between these groups is necessary for patient safety. PAH-specific therapies that are first-line for **Group 1** carry **Class III (Harm)** recommendations for **Groups 2 and 3**, which constitute the majority of pulmonary hypertension cases in the Medicare population (age >65).^{1,2}

WHO PH Classification

WHO GROUP	NAME	KEY ETIOLOGIES	TREATMENT APPROACH	PAH-SPECIFIC THERAPY?
Group 1	Pulmonary Arterial Hypertension (PAH)	Idiopathic, heritable, drug-induced, CTD-associated, HIV, portal hypertension, congenital heart disease	PAH-specific therapies: endothelin receptor antagonists, PDE5 inhibitors, prostacyclin pathway agents	Yes — first-line indication
Group 2	PH due to Left Heart Disease	LV systolic/diastolic dysfunction, valvular heart disease, congenital/acquired cardiovascular conditions	Treat underlying cardiac disease; diuretics; valve intervention if indicated	No — Class III (harm)
Group 3	PH due to Lung Diseases and/or Hypoxia	COPD, ILD (including IPF), sleep-disordered breathing, chronic high-altitude exposure	Treat underlying lung disease; supplemental oxygen; inhaled treprostinil (FDA-approved for PH-ILD)	No — Class III (harm); exception: inhaled treprostinil for PH-ILD
Group 4	Chronic Thromboembolic PH (CTEPH)	Organized thromboembolic obstruction of pulmonary arteries	Pulmonary thromboendarterectomy (potentially curative); balloon pulmonary angioplasty; riociguat for inoperable/residual disease	Riociguat only (not standard PAH therapies)

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